

Regular review

Diagnosis and management of porphyria

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Although porphyria is a relatively uncommon condition, it should be considered in patients presenting with an atypical medical, psychiatric, or surgical history. Acute attacks are associated with a substantial morbidity and mortality; there is a need for rapid and accurate diagnosis of the neuropsychiatric porphyrias, particularly because haem arginate can induce a definite remission if given early in an attack. Additionally, porphyrias may present with skin lesions or photosensitivity.

What are the porphyrias?

The porphyrias form a heterogeneous group of inherited disorders of haem biosynthesis, and they are often missed or wrongly diagnosed. A partial deficiency of one of the seven enzymes in the pathway causes characteristic clinical and biochemical features. These disorders are due to a specific alteration in the pattern of accumulation of porphyrin and porphyrin precursors (table). Each type of porphyria is defined by a unique pattern of accumulation and excretion of haem precursors, as well as a reduction in the relevant enzyme activity. Correct interpretation of the appropriate biochemical investigations is essential for accurately diagnosing and managing the porphyrias, as clinical features alone are not sufficiently specific either to confirm a diagnosis or to distinguish between the various forms.

There are seven main types of porphyria (fig 1), which are broadly classified according to clinical features into neuropsychiatric, dermatological, and mixed forms. Acute intermittent porphyria and plumboporphyria are predominantly neuropsychiatric; congenital erythropoietic porphyria, porphyria cutanea tarda, and erythropoietic protoporphyria have predominantly cutaneous manifestations; and hereditary coproporphyria and variegate porphyria are classified as mixed as they may have both cutaneous and neuropsychiatric features. The prevalence of porphyria varies widely from country to country and also depends on the type of porphyria. Overall prevalence of overt cases in the United Kingdom is about 1 in 25 000 population for porphyria cutanea tarda and less than 1 in one million for congenital erythropoietic porphyria.¹ Plumboporphyria has not been reported in Britain.

Summary points

The porphyrias form a group of inherited disorders of haem biosynthesis of which there are seven main types

Porphyrias can be classified into acute (neuropsychiatric), cutaneous, and mixed forms

Acute forms can be life threatening, but attacks can be aborted by early administration of haem arginate

The acute porphyrias are often misdiagnosed; most commonly they present as acute abdominal pain or as neurological or atypical psychiatric symptoms

Patients with porphyria should be referred to specialist centres and be advised to avoid precipitating factors, such as certain drugs

When a patient is diagnosed with an acute porphyria the whole family needs to be screened

Methods

We based this article on literature reviews, including a Medline search (1966-98), and on the many years' experience of our department in the management of the porphyrias. King's College Hospital is one of the two recently established supraregional assay service centres for laboratory diagnosis and for the provision of clinical advice on the management of porphyria.

General clinical aspects

Patients with porphyria present in three different ways—with cutaneous lesions, acute attacks (see below for features), or both. Clinically identical acute attacks can occur in acute intermittent porphyria, variegate porphyria, hereditary coproporphyria, and plumboporphyria.² Skin lesions accompany the acute attack in about half of patients with variegate porphyria and in about a third of patients with hereditary coproporphyria²; skin lesions may be the sole presentation in these porphyrias.

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Summary of diagnosis patterns of overproduction of haem precursors in different porphyrias

Type of porphyria	Urine concentration		Porphyrins in faeces	Porphyrins in erythrocytes
	Porphobilinogen and aminolaevulinic acid	Porphyrins		
Plumboporphyria*	Aminolaevulinic acid	Coproporphyrinogen III	Not increased	Zinc protoporphyrin
Acute intermittent porphyria	Porphobilinogen > aminolaevulinic acid	Porphyrin mainly from porphobilinogen	Normal, occasionally slight increase (coproporphyrinogen, protoporphyrin)	Not increased
Congenital erythropoietic porphyria	Not increased	Uroporphyrinogen I > coproporphyrinogen I	Coproporphyrinogen I	Zinc protoporphyrin, coproporphyrinogen, uroporphyrinogen
Porphyria cutanea tarda	Not increased	Uroporphyrinogen > heptacarboxylate	Isocoproporphyrinogen, heptacarboxylate	Not increased
Hereditary coproporphyria	Porphobilinogen > aminolaevulinic acid	Coproporphyrinogen III (porphyrin mainly from porphobilinogen)	Coproporphyrinogen III	Not increased
Variegate porphyria	Porphobilinogen > aminolaevulinic acid	Coproporphyrinogen III (porphyrin mainly from porphobilinogen)	Protoporphyrin IX > coproporphyrinogen III and porphyrin X	Not increased
Erythropoietic protoporphyria	Not increased	Not increased	Protoporphyrin increased or decreased	Protoporphyrin

*Lead poisoning produces an identical overproduction pattern.

Acute attacks

Acute intermittent porphyria is the commonest of the acute porphyrias. The clinical features of an acute attack vary greatly. The most common symptom is severe abdominal pain, which may be accompanied by neurological and psychiatric symptoms (fig 2).³ Muscular weakness, particularly a proximal myopathy affecting the arms, is common. Muscular weakness can, however, progress to quadraparesis and respiratory paralysis and arrest, which may resemble the Guillain-Barré syndrome. Mild sensory changes often accompany the predominantly motor neuropathy—often in a “bathing trunk” distribution.⁴

Marked constipation, nausea, vomiting, postural hypotension, and hypertension are other common features. Hyponatraemia occurs as a result of dehydration, nephrotoxicity, or occasionally inappropriate antidiuretic hormone secretion. The pathogenesis of the clinical features is poorly understood, but possible mechanisms include damage by free radicals,⁵ direct neurotoxicity of aminolaevulinic acid,⁶ and haem deficiency in nervous tissue.⁷ The recent development of

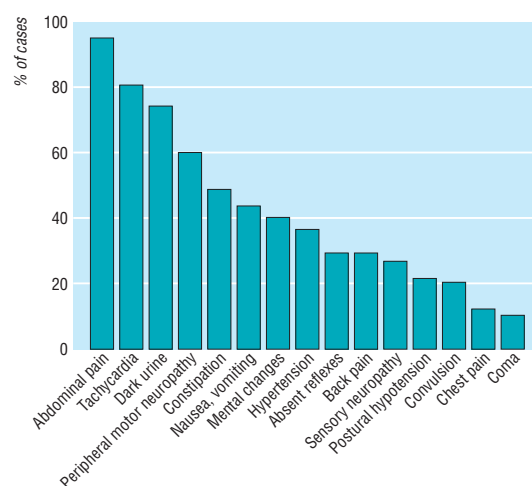


Fig 2 Frequency of signs and symptoms in acute porphyria. Adapted from Kappas et al² and McColl et al¹¹

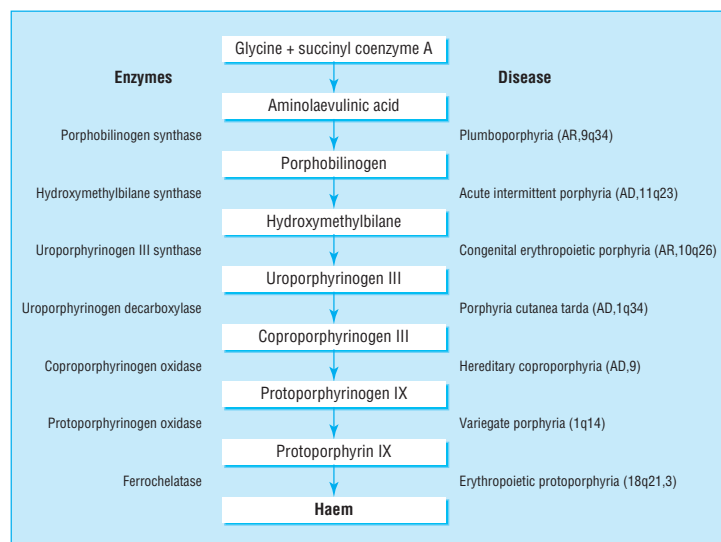


Fig 1 Pathway of haem synthesis. Blocks at various parts of the pathway result in different porphyrias (numbers in parentheses show inheritance). AR=autosomal recessive, AD=autosomal dominant

an animal model of gene knockout acute intermittent porphyria⁸ should allow rapid progress in this area.

The acute attacks are often triggered by exposure to exogenous precipitating factors,^{9,10} including a wide range of commonly prescribed drugs¹¹; illicit drugs including amphetamines, cocaine, and derivatives; alcohol misuse; fasting; stress; infection; sex hormone treatment; and smoking (box).¹² In women relapses occur particularly premenstrually and in pregnancy. Acute attacks are rare before puberty, are most common in people in their 30s, and are four to five times more common in females than in males, with a peak age of presentation in the early 30s. Only 10–15% of gene carriers develop the clinical syndrome. A third of patients have no family history, the condition probably having remained latent (inactive) or unidentified for several generations. The frequency and severity of the attacks vary widely. In some people the disease remains latent throughout life, even in the presence of precipitating factors. Other people experience frequent and sometimes life threatening attacks, even in the apparent absence of exogenous precipitating factors.

Factors that may precipitate acute attacks of porphyria

- Drugs—barbiturates and oestrogens (may be safe in replacement doses), progesterones, sulphonamides, methyl dopa, danazol, diazepam, phenytoin, carbamazepine, sulphonylureas, chloramphenicol, tetracyclines, some antihistamines
- Fasting
- Smoking
- Alcohol
- Substance misuse—particularly marijuana, ecstasy, amphetamines, and cocaine
- Infection
- Emotional and physical stress
- Cyclic factors—premenstrual attacks are common

Drug treatment should be prescribed only after reference to a drug list. Many such lists exist,^{11–13} but no list is universally accepted. Some drugs are generally agreed to be safe in acute porphyria; some are considered to be unsafe; and a large number of drugs may or may not be safe. For this last group of drugs, a commonsense assessment of benefit versus risk is needed; an acute attack is less likely to be precipitated if the disease is latent, if the patient has previously only had a single attack, and if the concentrations of urinary porphobilinogen and particularly of aminolaevulinic acid are normal at the time of prescribing the drug.

Laboratory diagnosis

When a porphyric patient presents with symptoms of a possible acute attack, the key question is whether these are due to porphyria; not all symptoms in porphyric patients are due to porphyria—porphyric patients are not immune to other conditions.

Acute attacks of porphyria are invariably associated with raised urinary excretion of aminolaevulinic acid and porphobilinogen. A fresh urine sample, protected from light, should be sent to a specialist laboratory for accurate quantitation of aminolaevulinic acid and porphobilinogen concentrations. Routine laboratory screening tests may be unreliable,¹⁴ but a recently introduced screening kit seems promising.¹⁵ A clue to the diagnosis is that the urine is often dark on standing owing to polymerisation of porphobilinogen to porphyrins and other pigments. Between attacks, however, concentrations of urinary porphobilinogen and particularly aminolaevulinic acid are often normal. Plasma fluorescence is usually increased in variegate porphyria and is valuable both diagnostically and for family studies.^{16–17}

Acute intermittent porphyria, variegate porphyria, and hereditary coproporphyria are differentiated by analysis of faecal porphyrins (table). The diagnosis of acute intermittent porphyria can be further confirmed by the demonstration of reduced red cell hydroxymethylbilane synthase activity,² although it is important to check that routine haematological indices are normal. DNA analysis is not yet of routine diagnostic value.

Management of acute attacks

About 1% of acute attacks of porphyria may be fatal. Most patients with an acute attack will require

admission to hospital. Only drugs known to be safe in porphyria should be prescribed. For severe pain, opiates are safe—that is, non-porphyrigenic. Pethidine, morphine, or diamorphine can be given. Chlorpromazine may be helpful to promote relaxation and sleep. Sympathetic overactivity causing tachycardia and hypertension can be alleviated with propranolol.

Convulsions may occur during an acute attack. Their onset may be precipitated by hyponatraemia, so plasma osmolality and electrolyte values should always be checked.² Treatment is by fluid restriction, and the convulsions usually resolve as the attack subsides. Rarely, epilepsy may be the presenting symptom in acute intermittent porphyria. Treatment should be aimed at the underlying disease. Seizures are notoriously hard to manage in porphyria as all commonly used anticonvulsant agents are porphyrigenic and may therefore lead to further exacerbation of the disease.^{18–19}

Historically, bromides have been the drug of choice for seizure control in most patients with acute porphyria.^{18–20} However, in the mid to late 1990s several new antiepileptic drugs were introduced, and, of these, gabapentin^{21–23} and vigabatrin²¹ were shown to be successful in seizure control without inducing porphyric crises. These drugs are now the treatment of choice for these patients.

Oral and intravenous glucose (for maintaining a high energy intake) and haem arginate are the mainstay of treatment. They reduce synthesis of aminolaevulinic acid, resulting in a clinical and biochemical remission, with urinary excretion of aminolaevulinic acid and porphobilinogen falling towards normal values. Glucose (10% intravenously) should be given for mild attacks and pending administration of haem arginate.

Haem arginate (Normosang, Leiras Medica, Finland) should be given at an early stage in an attack²⁴ as it cannot reverse an established neuropathy. It is given at a dose of 3 mg/kg/day for 4 days over 15 minutes by slow intravenous infusion. It is highly irritant so should be given by a central venous line. It acts rapidly and has a dramatic effect within one week. It is expensive and is available on an urgent need basis from the company (Orphan Europe, Henley-on-Thames, Oxfordshire) or from the two supraregional assay service centres. Recently tin protoporphyrin, an inhibitor of haem oxygenase, has been shown to prolong remission when given with haem arginate.²⁵ Its side effects of cutaneous photosensitivity and potential toxicity limit its use, however, and further evaluation is continuing. A small proportion of patients—mainly females—have recurrent attacks with no apparent precipitants. Prophylactic haem arginate may be necessary on a regular basis for such patients. Induction of a chemical menopause with luteinising hormone releasing hormone agonists has been successfully used in this situation.

Prevention of attacks

Precipitating factors mentioned previously (such as alcohol, smoking, and porphyrigenic agents) should be avoided, as should sudden or prolonged low energy diets. Patients should wear a Medic Alert bracelet (an emergency identification bracelet) and should be fully educated regarding precipitating factors. They should be given an information booklet, be encouraged to join



Fig 3 Cutaneous lesions in porphyria cutanea tarda

a support group—for example, the British Porphyria Association (14 Mollison Rise, Gravesend, Kent DA12 4QJ)—and be referred to a specialist centre. Attacks may occur during pregnancy, when oestrogen concentrations are high, and the patient should be advised to avoid pregnancy until she has been in remission for at least two years. Aminolaevulinic acid can cross the placenta and possibly cause toxicity to the developing fetal brain.²⁶ If acute attacks do occur during pregnancy they should be treated in the normal way. Haem arginate has been used successfully in pregnancy.

Cutaneous porphyrias

Photosensitisation is due to accumulation of porphyrins in the skin and may present as one of two distinct syndromes. Erythropoietic protoporphyria presents in childhood with acute photosensitivity—that is, burning, itching, and erythema on exposure to sunlight, but without bullae and with minimal scarring—and is due to an inherited partial deficiency of ferrochelatase.²⁷ The disease usually presents in childhood, and there is also accumulation of protoporphyrin in the liver leading to cholelithiasis and occasionally liver failure.

Skin lesions in the remaining cutaneous porphyrias (porphyria cutanea tarda, variegate porphyria, hereditary coproporphyria, and congenital erythropoietic porphyria) are similar and include fragile skin that heals slowly after minor trauma, subepidermal bullae (fig 3), pigmentation, and hypertrichosis, particularly on the forehead and upper cheeks. The lesions occur in skin exposed to the sun and are most severe in congenital erythropoietic porphyria.²⁸ Porphyria cutanea tarda is the commonest of all the porphyrias (table). Most cases are associated with liver cell damage. There are two forms.

Sporadic porphyria cutanea tarda—The sporadic form accounts for 80-90% of all cases of porphyria cutanea tarda. Aetiological factors include alcohol, oestrogens, iron, and chemicals (for example, hexachlorobenzene). There is also an association with hepatitis C. Mild iron overload is nearly always present. People with genetic haemochromatosis are four times more likely than otherwise normal subjects to have sporadic porphyria cutanea tarda,²⁹ and these patients need to be investigated for iron overload—for example, ferritin

and liver iron concentrations and DNA studies for the coexistence of genetic haemochromatosis.

Familial porphyria cutanea tarda—The familial form accounts for 10-20% of all cases of porphyria cutanea tarda. Autosomal dominant hepatoerythropoietic porphyria (a severe rare homozygous form) also exists, but environmental factors are also important in the expression of the disease. Biochemically, minor rises in liver toxicity and ferritin concentrations are common. Liver biopsy usually shows fatty infiltration and inflammatory changes, with cirrhosis being present in a third of cases.³⁰ Porphyria cutanea tarda increases the risk of hepatocellular carcinoma in patients with chronic liver disease.³⁰⁻³² Treatment of the contributory hepatotoxic agents—for example, abstinence for alcohol misuse, venesection for iron overload, and, somewhat controversially, chloroquine therapy—can lead to a useful improvement in the skin lesions, with long term remission in some patients.³³ Chloroquine forms a complex with uroporphyrin and promotes release of uroporphyrin from the liver; it may also inhibit the synthesis of uroporphyrin.³⁴ Use of ultraviolet blockers is also valuable.

Laboratory diagnosis of cutaneous porphyrias

Erythropoietic protoporphyria is diagnosed or excluded by measuring erythrocyte free protoporphyrin. Analysis of urine and faeces is helpful (table).

In patients who are suspected of having one of the remaining cutaneous porphyrias it is essential to examine both urine and faeces for excess porphyrins as the water solubility (and hence route of excretion) of the individual porphyrins differs. Positive findings should be followed up by fractionation to identify and quantitate the individual porphyrins (including isomers). This will usually entail referral to a specialist laboratory with experience of the porphyrias. It is particularly important to distinguish between those patients with variegate porphyria and hereditary coproporphyria presenting with skin lesions alone (who will be at risk of from life threatening acute neurological attacks if exposed to precipitants) and those with porphyria cutanea tarda or congenital erythropoietic porphyria (who are not at risk from such attacks).

Management of cutaneous porphyrias

The cutaneous porphyrias are treated by avoidance of sunlight and attention to skin care. Additionally, venesection to deplete excess iron stores, oral chloroquine to increase urinary porphyrin excretion, and avoidance of alcohol and oestrogens are all particularly helpful in porphyria cutanea tarda. In erythropoietic protoporphyria, in addition to the above, administration of carotene³⁵ is often helpful and is believed to work by quenching active oxygen species.

Screening and family studies

The dominant mode of inheritance of the acute porphyrias, the occurrence of asymptomatic gene carriers, and the risk of developing potentially fatal attacks of neuropsychiatric origin if someone is exposed to a wide range of common precipitating factors make it essential to exclude or confirm the diagnosis of neuropsychiatric porphyria in all relatives whenever this has been diagnosed in any family member. In some cases, however, because of the equivocal

biochemical analyses, confirming or refuting the inheritance of acute porphyria is not always possible. In these cases, genetic analysis is increasingly used, but this is sometimes difficult. In acute intermittent porphyria, for example, there are over 100 mutations including deletions, insertions, and missense, non-sense, and splicing mutations, and most of these mutations are family specific.³⁶ However, the gene locations of many of the porphyrias have been identified,^{1 11} and rapid progress is expected in this area.

Clearly, families benefit when the mutation is detected. Not only will detection identify members with latent porphyria, but also unaffected members can be distinguished with confidence.³⁶ Exclusion of carrier status makes it unnecessary to follow restrictions to prevent attacks and avoids the need for screening of future generations of that particular family branch.

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Why did I become a doctor?

A wise headmaster

The acknowledged importance of career advice and guidance has led to a burgeoning cottage industry with the involvement of numerous stakeholders. Starting at school and continuing through the undergraduate and postgraduate phases, increasingly sophisticated techniques are being used to try to avoid square pegs ending up in round holes. However, an effective directional steer can sometimes be given by a person with the ability to see the big picture.

Career advice at my school was the province of the headmaster, who saw each pupil individually in the fourth year. A dour Scot, he was of the no pussyfooting genre, having previously been a house master at a school in Scotland with a justifiable reputation for its spartan lifestyle. I duly reported to the headmaster's study at the appointed time, having carefully rehearsed my piece. Like most teenagers I was anxious to leave school at the earliest opportunity and earn some money in the big wide world.

"Well Salter, what do you want to do?" he asked. "I would like to leave after O levels and go into either banking or pharmacy," I

said. At that time neither of these required A levels or a degree. His career guidance was short and to the point. "Och Salter, you're staying on to the sixth form," he said. The interview was promptly terminated, and I was shown the door.

Fortunately, those were the days when authority, particularly the headmaster's, was never challenged. I stayed, opted for medicine, and have thoroughly enjoyed my career as a consultant physician and clinical tutor. I have remained eternally grateful for such short but effective career guidance.

R H Salter *clinical tutor, Carlisle*

We welcome articles of up to 600 words on topics such as A memorable patient, A paper that changed my practice, My most unfortunate mistake, or any other piece conveying instruction, pathos, or humour. If possible the article should be supplied on a disk. Permission is needed from the patient or a relative if an identifiable patient is referred to.